ELLIOT (G. T.)

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A CASE OF DERMATITIS HERPETIFORMIS.

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THE following case of dermatitis herpetiformis is so typical in its entire clinical symptomatology and course that it would appear worthy of record. The repeated and constant report of similar instances of cutaneous disease has in the last few years greatly increased our knowledge of the general features which characterize it, as well as broadened our conception of its nature. At the same time, the diagnostic value of its symptoms is more fully appreciated, the disease is more readily and generally recognized, and no longer confounded with those one and other cutaneous processes which at various times in its course it may resemble, owing to the individual lesions present.

Male, aged fifty-six, a merchant, consulted me February 1, 1890. He had never been ill, and his domestic and business relations were entirely free from any such complications as would produce worry, anxiety, or lead to any moral or other shocks. Temperate, a moderate smoker and not given to any excesses, except indulgence in strawberries whenever obtainable, he could give no clew in his history suggesting the origin of his disease. The first outbreak was preceded for a year or more by a gradual dryness, harshness, and roughness of the entire skin, but especially of the hands, and in June, 1889, an intensely itchy eruption appeared suddenly over his chest. It was thought to be "prickly heat," but, as it spread rapidly over the back and extremities, a physician was consulted. At that time, the lesions were papules and vesicles grouped and arranged in rings, and the diagnosis was made of "ringworm" complicated with eczema. The application of the remedies ordered led to extensive exfoliation of the epidermis, and immediately afterward, bullæ as large as a pigeon's egg appeared around the wrists, in the axillæ and in the groins. Similar lesions, pea to hazelnut in size, developed in crops generally over the body, except upon the back. During the succeeding six months, the erup-

tion persisted almost continuously, though the type of lesion was not stated, relapse succeeding relapse, each separated by only short intervals of improvement. Intense, agonizing itching was present all the time, as well as a severe universal hyperidrosis, and violent clonic spasms of the extremities caused him much distress when lying down. By December of the same year his condition had considerably improved, and he went to Old Point Comfort. During the first two weeks he gained in strength, he slept better, the itching and sweating diminished, but he then received accidentally a severe moral shock. The following day, the pruritus returned and bullæ cropped out on the inner surfaces of the thighs. A few days later, papules appeared over the extremities, and the outbreak became progressively severer—papules, vesicles, bullæ, and erythematous patches developing in successive crops over the various portions of the body. Shortly after, he consulted Dr. Duhring in Philadelphia, who diagnosed the case as one of dermatitis herpetiformis, and later he came to New York and under my care.

On examination, the patient was found to be well nourished, his appetite and digestion were good, but the bowels for several months had been loose, three to four or more stools occurring every day. From two and a half to four quarts of urine were passed every twenty-four hours, frequent nocturnal micturition being a necessity. The amount would be increased whenever the itching was severe, or the patient became worried or depressed. The urine was limpid and very light in color, acid, and of a specific gravity of 1·019. No albumin or sugar. Urea, 1·60 per cent. Vesical and urethral epithelium, a few pus cells and small uric-acid crystals were found present under the microscope. No elevation of the temperature existed or took place while under treatment. The pulse was full and regular, but when he exerted himself one beat in about fifteen would be missed—a condition which had existed for twenty-five years. As far as could be ascertained, all the other organs of the body were normal. The mind was clear, the brain active, no muscular weakness, no tenderness on pressure over the spine. The hair had turned very gray since the disease existed, but there was no alopecia. The beard had changed from brown to white. The nails of the fingers and toes were dystrophic, discolored, thickened and split longitudinally, and their growth was apparently retarded.

With the exception of the face, the entire body was deeply pigmented, but here and there were small islands of skin normal in color, and standing out in vivid contrast with the dark brown left by former outbreaks, with the red hue of the newer erythematous lesions and

with the purplish and cyanotic tinge of those undergoing involution. On the upper arms and on the abdomen were many dirty, grayish-white, irregularly shaped spots, evidently superficial cicatrices caused by the scratching and wounding of the lesions. The skin in its entirety was much thickened and indurated, especially over the buttocks. Below the knees it was tense and shiny, though not œdematous. The lesions distributed over the surface were of all kinds, forms, shapes, and sizes. Vesicles, bullæ, papules, and erythematous bands and patches occurred alone on a surface, or were grouped together in a most peculiar and inextricable manner. On the face, bullæ alone were present. They were discrete or grouped, the size of a buckshot to a hazelnut, round or oval, linear or crescentic, or curving partly around a central bleb. Some were angular and stellate, others of uniform outline.

Over the body, discrete bullæ were distributed. They were tense, resistant, not rupturing easily, and contained a clear, light-yellow, neutral fluid. In some places, notably at the navel, a large central bulla was seen surrounded by a row of vesicles somewhat larger than a pinhead. These bullous lesions arose apparently from the skin, without being preceded by any redness or primary change. The vesicles were pinhead and a little larger in size and grouped together, the groups in some places being a handbreadth in size. Some were deep-seated, as though occupying the center of pre-existing papules. Only on the backs of the hands had the contents of either bullæ or vesicles become purulent, and when uninjured by scratching they dried up into thin, yellowish crusts. The papules varied in size from a small pea to a pinhead, bright or dusky red in color, and resistant. The larger ones were discrete, but the others were arranged in groups occupying small or large areas. The erythematous lesions were of most varied shape and form, and ranged in size from that of a nickel to patches occupying, for instance, an entire buttock. They were also represented by narrow, elevated bands, ring-shaped and inclosing a sunken-in, violaceous center, or by segments of circles, or by gyrate or festooned lesions of varying length. In their evolution, they began as an erythematous solid efflorescence about the size of a nickel; the central portion soon subsided, becoming dusky red or cyanotic in color, while the red border, a quarter to a third of an inch broad, advanced. As progression took place, involution occurred *pari passu*, until a limit of growth seemed to be reached, and then the entire lesion disappeared, leaving only some pigmentation, or upon the red elevated margin a row of vesicles or papulo-vesicles developed. The lesion would then appear as made up of a dusky-red or brownish disk, entirely or partly encircled

by a chaplet of distinct vesicles arranged in a uniform or in a more or less irregular manner. Separate vesicles never arose in the centers of these disks, nor were these latter encircled by more than one row of such lesions. Instead of the progressive enlargement taking place, sometimes disks appeared which, from the first, were surrounded by a vesicular border. There were also crescentic lesions and others representing some segment of a circle which appeared as primary efflorescences. In their advance over the surface they would join together and thus form gyrate and wreathlike figures of varying contour and extent. On the legs below the knees there were many excoriated places and superficial ulcerations, due to scratching and the tearing of the surface.

No particular course was followed in the evolution or involution of the lesions. They would appear suddenly, singly or in crops, and remain for several days, or they would come out and disappear in a few hours or in a day. These latter would leave no trace of their former presence, but the bullæ or vesicles would dry up into crusts, or, being ruptured by scratching, leave an excoriated surface which healed slowly. The papular and erythematous lesions, however, would change from a bright to a dusky red, then purplish, and when entirely gone only a brown pigmented area remained.

The subjective symptoms were intensely severe, and consisted entirely in intolerable itching and burning. In some degree they were always present, but there would be paroxysmal exacerbations of excessive severity, and more frequently at night, or whenever he removed his clothing and his skin became exposed to the air. These symptoms and the clonic spasms already referred to prevented sleep for some hours after he went to bed. The itching and burning would also become worse whenever the weather was stormy or rainy, or when the bowels were costive, or if any mental worry or anxiety supervened. Before the appearance of a crop of lesions, the sites upon which they were to appear would likewise itch and burn intensely. There was a universal severe hyperidrosis also present, which added to his discomfort. Always existing on the trunk, it became severe upon the extremities only during the pruritic paroxysms.

While the patient was under treatment, there was progressive improvement, interrupted, however, by a succession of relapses of varying extent and intensity, and consisting of a single type of lesion or of multiform efflorescences. The occurrence of a relapse could not always be traced to any definite cause, though it would take place when there was a change from fine to bad weather, when any functional disturbance of the gastro-intestinal canal arose, or when he

overtaxed himself mentally or physically. Gradually, however, these renewed outbreaks became less frequent and severe, the clonic spasms as well as the hyperidrosis ceased, the pigmentation diminished, the skin was less thickened and infiltrated, and, generally and markedly improved physically and mentally, the patient went home March 23d. He was then entirely free from any lesions, and had only occasionally a little itching. At the end of a few weeks, however, the cutaneous symptoms returned in some degree, but subsided shortly. In the following year there were several relapses, and finally he went to Europe for a number of months, and has since then remained comparatively well. The treatment he received while under my care consisted of general dietetic and hygienic measures. Ergot internally up to a drachm and a half three times a day, and ichthyol in carron oil externally. Besides these, baths of starch and white-oak bark were given daily. The ichthyol gave almost instantaneous relief to the burning and itching, and enabled him to obtain rest and sleep.

As I have already stated, this case presented in a most typical manner all the symptoms characterizing dermatitis herpetiformis, and in this it agreed with the many other examples of the disease which I have had under my care. That at times during its course it resembled to some extent in its lesions other diseases of the skin, is unquestionable—in fact, that is true in all cases of dermatitis herpetiformis—yet when the symptoms developed were taken as a whole, it was readily seen that each variation in aspect was simply an expression of a part of that pathological process now recognized and known in its variable and multiform phenomena as Duhring's disease. The right of distinct existence is not, however, accorded to dermatitis herpetiformis by all, as only in the last year Kaposi * has denied that it was a disease of itself, but rather a sort of collective conception, made up in part of erythema multiforme and of other bullous affections, urticaria, etc., and in part of pemphigus. As far as pemphigus is concerned, I most assuredly can affirm that this patient was not affected with that disease. There were at various times during its course bullæ on the surface, it is true, but these were greatly in the minority; they were distributed here and there, but were relative exceptions in the great mass of other lesions with which the surface was covered. Papules and vesicles arranged in groups were the predominant lesions, as well as erythematous patches of various shapes. I can not see how a pemphigus could be made out of such a case, nor how its clinical history, phenomena, behavior, or course could be reconciled with those of pem-

* Proceedings of Fifth German Dermatol. Congress.

phigus as defined and described by authors. Pemphigus acutus, a severe disease, with systemic reaction, fever, etc., and usually ending fatally in a few days, does not come into consideration here, nor does pemphigus foliaceus or vegetans, but only pemphigus vulgaris chronicus. This disease is defined by Hebra* as characterized by repeated outbreaks of *bullæ*, and no other lesion is described or mentioned by him in the whole article. Erasmus Wilson,† Tilbury Fox,‡ Hardy,# Simon,‖ Crocker,△ Brocq,◊ Zeisler,‡ and any number of others, have not deviated from this, Hebra's definition, beyond mentioning the occasional presence of some erythematous patches or urticarial wheals upon which bullæ subsequently developed. Kaposi‡ himself, in his book (1883), and again in its newer edition (1893), defines pemphigus as a "bullous eruption, characterized by outbreaks of bullæ," mentioning that erythematous phenomena of various kinds accompany it. No other lesions are described, and in the article devoted to the subject he agrees with those other writers mentioned. When the description of pemphigus as obtained from the works of these eminent writers is compared with that of dermatitis herpetiformis as given by Duhring, Brocq, and others, and also as observed by myself in many cases, it is certainly difficult to understand a failure to recognize Duhring's disease as distinct from pemphigus: on the one hand, a process of which the essential symptoms—the *sine qua non*—are bullæ; on the other hand, a disease in which bullæ do occur, but only as one of many other types of lesions, and, moreover, one which in my experience at least, is more uncommon than any other of the eruptive phenomena seen in the process. Besides these, in dermatitis herpetiformis the subjective symptoms are intense, the itching, burning, and paræsthesia always severe; in pemphigus they are very slight, and often entirely wanting. The latter disease is accompanied by severe systemic symptoms, the patients become emaciated and prostrated, while in the former the individual is in a relatively good condition of health, even though the process runs a long and chronic course. The mortality of pemphigus is certainly very high, and it has always been considered a serious disease; that of dermatitis herpetiformis is infini-

* Hebra. Hebra and Kaposi, *Hautkrankh.*, 1872.
† Wilson. *Diseases of the Skin*, 1867.
‡ T. Fox. *Skin Diseases*, 1873.
# Hardy. *Traité des maladies de la peau*, 1886.
‖ Simon. *Hauthkrankheiten*, 1851.
△ Crocker. *Diseases of the Skin*, 1888.
◊ Brocq. *Traitement des maladies de la peau*, 1890.
‡ Zeisler. Morrow's *System of Syph.*, etc., 1894.
‡ Kaposi. *Pathologie und Therapie d. Hautk.*, 1883 and 1893.

tesimal. I do not remember more than one case of pemphigus vulgaris which I have seen, which has not ended fatally, while in over forty of dermatitis herpetiformis which I have treated only one died, and then from septicæmia, the origin of which was very doubtful.*

In view of these striking differences between the two forms of eruption, differences which have repeatedly been stated, it is therefore not surprising that the consensus of opinion has accepted and substantiated Duhring's description, and that dermatitis herpetiformis has become recognized, with few exceptions, as a distinct entity. It may, however, be advanced that such cases are nevertheless examples of pemphigus vulgaris, being of the type pemphigus pruriginosus. Still, this latter is now separated from pemphigus, and certainly clinically it bears no resemblance to it. Kaposi† specifically states that the picture presented by pemphigus pruriginosus is entirely different from the usual one of pemphigus vulgaris (*ein von dem gewöhnlichen ganz abweichendes Krankheitsbild*), explaining the absence of bullæ by the fact that the erythematous lesions are scratched before the bullæ can form. I most certainly would agree with him that it bears no resemblance to pemphigus vulgaris, but, being so markedly representative of dermatitis herpetiformis, it is not surprising that it has come to be generally regarded as belonging in this latter category.

It is hardly necessary to make a differential diagnosis between this case of dermatitis herpetiformis and erythema multiforme bullosum, or of other type, or to take urticaria bullosum into consideration. Only careless observation could allow these eruptions to be confounded with the former one; and I would only state, in conclusion, that the case reported here was neither a pemphigus, nor an erythema multiforme, nor an urticaria, but a most typical and characteristic example of dermatitis herpetiformis.

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* Elliot. *American Journal of the Medical Sciences*, 1895. † Kaposi, *loc. cit.*